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# Bone Lesions in Children with Neurofibromatosis

*Nikolaos Laliotis*

## Abstract

Neurofibromatosis is often related with severe orthopaedic disorders in children. Bone lesions are rare but pose severe difficulties in management. It affects the spine and long bones. Lesions are associated either from enlargement of neurofibromas that affect the normal growth or from primary neurofibromatosis of long bones. Dystrophic scoliosis appears with short curves, with kyphosis and rotation of the apical vertebrae. Usually affect the thoracic spine, with penciling of the ribs. Surgical treatment is challenging in cases of rapid progression. Scoliosis may appear with curvatures similar to those in idiopathic scoliosis, without dysplastic changes of the vertebrae. Anterior bowing of the tibia is manifestation of NF and is distinguished from the benign posterolateral bowing. Evaluation of the medullary canal and presence of cystic lesions in the tibia is essential. Progression to pseudoarthrosis or pathologic fracture is common. Surgical management of tibial pseudoarthrosis remains a difficult procedure. Pseudoarthrosis may appear in fibula, radius or ulna but are extremely rare. Irregular eccentric bone cysts in long bones that are commonly diagnosed after a pathologic fracture, must be differentiated for NF. Malignant transformation of neurofibromas must be considered when there is rapid progression of the lesion.

**Keywords:** Scoliosis, dystrophic scoliosis, surgical management, spine in neurofibromatosis, spinal instrumentation, Congenital pseudoarthrosis tibia, fibula, radius, ulna, Idiopathic non-union tibia, fibula, radius, ulna, Neurofibromatosis tibia, fibula, radius, ulna

## 1. Introduction

Neurofibromatosis is a hereditary autosomal dominant disease associated with abnormal increase of neural cells, both from the central and peripheral nervous system. Children and adults are affected from the disease.

Orthopaedic manifestations of NF in children are found in the spine and the long bones. Alterations of the normal shape of the spine both in the frontal and sagittal plane appear, in the form of dystrophic and non-dystrophic scoliosis and kyphosis. It is unclear the exact mechanism for development of scoliosis is NF, as in general for scoliosis. Vertebral neurofibromas can erode the vertebrae either from the interior or the exterior, resembling congenital hemivertebra. Vascular and osteoblastic dysfunction may alter the shape of the vertebrae. Diagnosis of NF is based on the clinical criteria that include the dysplasia of long bones and spine lesions. Radiological evaluation both of x-rays and MRI and is important to properly follow the affected children. Management of dystrophic curves is a challenge for the pediatric spine surgeon.

Neurofibromatosis lesions of long bones are extremely rare, affecting usually the tibia and fibula in the lower limbs and radius and ulna in the upper limb. They appear with the form of congenital pseudoarthrosis. Treatment requires expertise since complications and relapse are not uncommon.

## 2. Spine lesions in neurofibromatosis

### 2.1 Scoliosis

Scoliosis is the most common osseous involvement in NF.

The incidence of scoliosis among children with NF is increased and it is referred that 10–60% of NF patients present with some type of spinal deformity. Approximately 3% of patients referred for scoliosis, have NF-1.

There are two patterns of scoliosis in neurofibromatosis: dysplastic and non-dysplastic.

Scoliosis appears early in life of children with NF, usually at the age of 5–8 years old. They belong to early onset scoliosis.

The dysplastic scoliotic curve is a short rigid curve with sharp angulation, usually located in the thoracic area. This is the primary curve. As the patient grows, scoliosis may involve the cervical or the thoracolumbar spine. Scoliosis in the lumbar spine is rare. The apical vertebra appears with wedging, there is scalloping of the vertebral bodies and pencilling of the apical ribs. This wedging is resembling an hemivertebra. The foramen is enlarged and neurofibroma may be found entering the canal. They present with simultaneous kyphosis, with a sharp angulation at the apex of the curvature. These curves have the tendency to progress rapidly and often require early fusion to prevent the progression of the scoliosis [1–5].

Bracing has limited success to prevent progression of the curvature. For dystrophic curves greater than 40°, posterior spinal fusion with segmental instrumentation is the procedure of choice. It is important to use autologous bone graft, to enhance solid fusion. Solid curves, with kyphosis less than 40° angle, can be managed with posterior fusion, while with the presence of severe kyphosis concomitant anterior fusion is required. Despite solid fusion, some curves continue to progress. In severe kyphoscoliosis curves, both anterior and posterior procedures are required to achieve stability. It is important to use intra operative neurophysiological monitoring for these patients.

Pedicle morphology of dystrophic curves have differences compared with those in idiopathic scoliosis. There are often abnormal pedicles that result in misplacement of pedicles screws. Using CT measurements, abnormal types of pedicles are significantly more common in NF-1 scoliosis. A 3 D navigation system was used for accurate placement of screws. In proximal thoracic pedicles, with small diameter, sublaminar hooks may be used [6–9].

Management of severe dystrophic curves is a challenge for the paediatric spine surgeon. Early fusion may interfere with the body development, lung and pulmonary function. The use of growing rods (GR) that can correct deformities, without fusion, become now the standard procedure for management of early onset scoliosis. They require periodically lengthening [10–12]. Carbone et al. [10] in a series of 7 children with dystrophic thoracic scoliosis, report the results from the use of GR. They achieved a scoliosis correction measuring the Cobb angle from 82,7° to 46,6°. Despite that they report 12 complications in 4 patients (47%), including rod breakage, their results are very promising for the management of the severe problem. They were lengthening the rods in a year basis program.

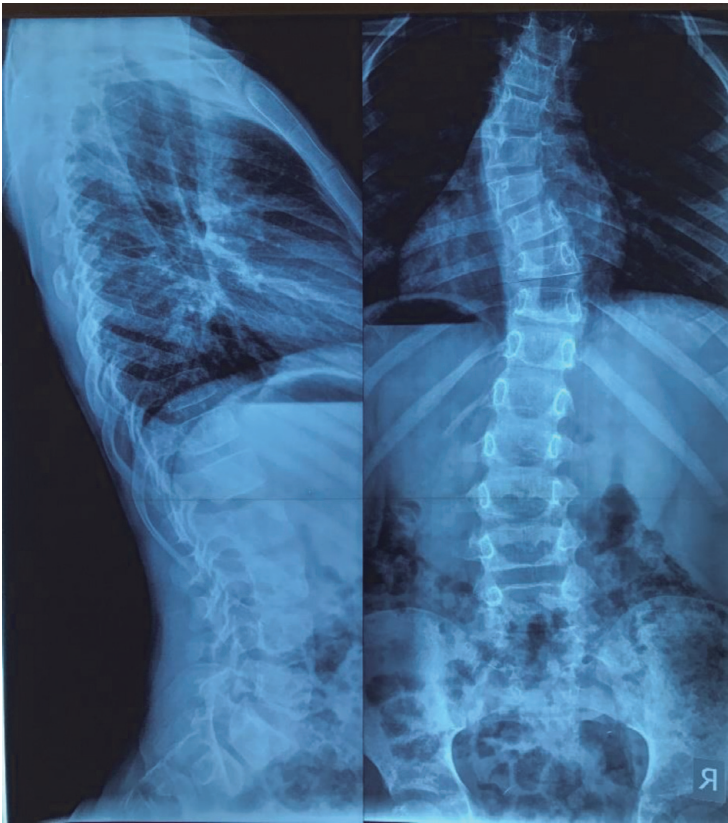
Yao et al. [13] compared the use of initial fusion in dystrophic curves, with the use of GR. Fusion achieved better correction and the complication rate reported was

lower in those treated with fusion. They report the technical difficulties to use fixation device because of the pedicle deformity. It is important to include all dystrophic vertebrae in the fusion area, to minimize complications.

High incidence of pseudoarthrosis in dystrophic curves have been reported, rising up to 60% in older reports [1]. Complication rate after surgery in children with dystrophic curves is expected to be higher than those surgically treated for idiopathic scoliosis [13, 14]. But recently, with the improvement of the procedures, Lyu et al. [15] report similar good results for both groups, with no fusion failure and similar rate of complications between the 2 groups.

Paraplegia may appear from the erosion of the vertebral bodies from neurofibromatosis tissue. It is of great importance to exclude the possibility of rib progression as a cause of paraplegia. CT scan and MRI are helpful to diagnose intraspinal rib progression. Resection of the rib as the first step, permit adequate correction of the curve with instrumentation, without risk for neurological deficit. In flexible curves, traction may result in reversal of the neurological deficit. Improvement of Frankel scores and rotatory thoracic subluxation using pre- operative halo traction has been reported. Traction (Halo traction) has no effect on rigid curves [12, 16, 17].

Non dysplastic scoliosis has similar natural history to idiopathic scoliosis that is seen in adolescents. They progress similarly to idiopathic scoliosis. We consider the balance of the body and the aesthetic shape of the body, as important parameters for management. Scoliosis with Cobb angle less than  $20^{\circ}$  is only observed. Curves between  $20$  and  $35^{\circ}$  may be managed with appropriate braces. Greater curves may require surgical correction and stabilization. It is important to regularly monitor these non-dysplastic curves, as they may become dysplastic with sudden increase of the curvature. This phenomenon of modulation, was first reported by Durrani [18]. Rib pencilling is an early sign for severe progression. MRI examination of the spine, every year is essential for the evaluation of the cord condition in NF scoliosis (Figures 1–3).

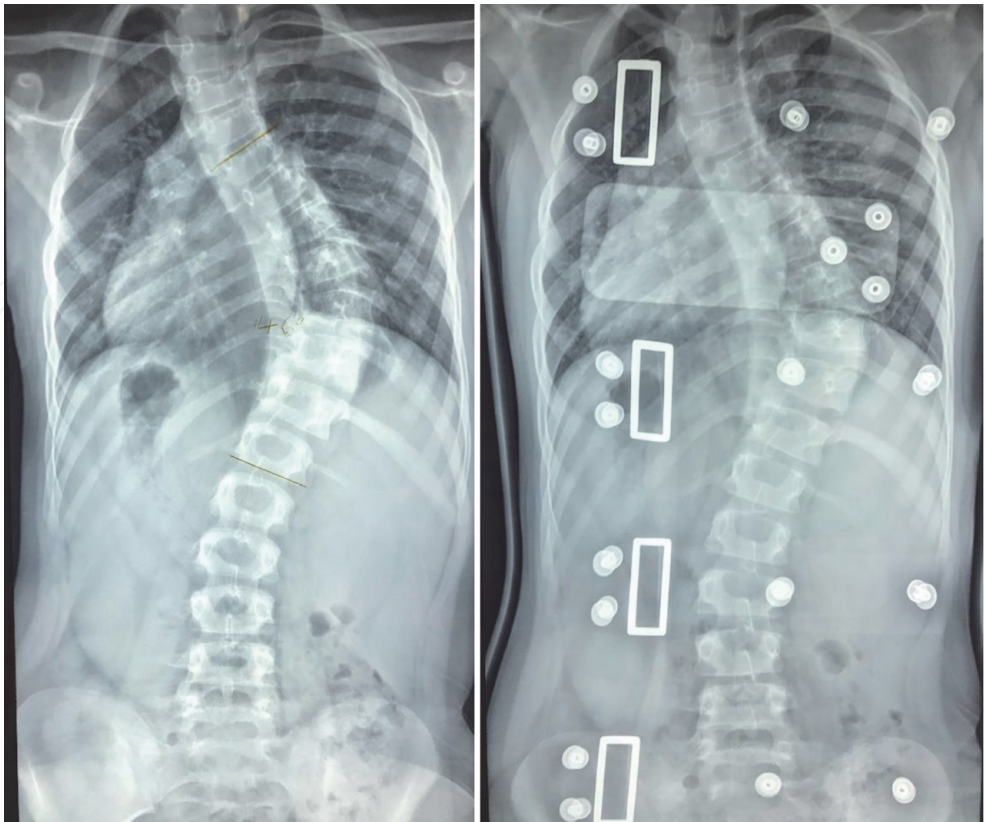


**Figure 1.**  
*Xray of non-dysplastic left thoracic scoliosis, in a boy with NF-1.*





**Figure 2.**  
*Photo of the body (anterior and posterior) of a 13 years old boy with right thoracic scoliosis and skin manifestations of NF.*



**Figure 3.**  
*Xray shows a non dystrophic right thoracic scoliosis. Brace treatment was ineffective to regress progression. He was advised for surgical correction.*

## 2.2 Cervical spinal lesions in NF

Cervical spine scoliosis is usually present with the sharp short kyphoscoliosis lesions of the thoracic spine. They may be found early in life.

Atlanto axial dislocations are reported in NF. Lesions of the upper cervical vertebrae, scalloping and erosion of the dens either from the eroding neurofibromas or from the pressure of the dural ectasia, are found. Children that had previously surgically treated for a neck mass, must be followed for recurrence and extension of the lesion in the upper cervical spine.

Patients may present neurological deficit. Prevention requires appropriate spinal instrumentation for stability and avoid of laminectomies. Patients with cervical spine involvement may present with torticollis or dysphagia. Manifestations of vertebral cervical involvement are found in radiological evaluation, with scalloping of the vertebrae, enlargement of the foramen due to neurofibroma. It is important to perform x-ray evaluation of the cervical spine for patients with NF that are scheduled for general anaesthesia.

## 2.3 Spondylolisthesis

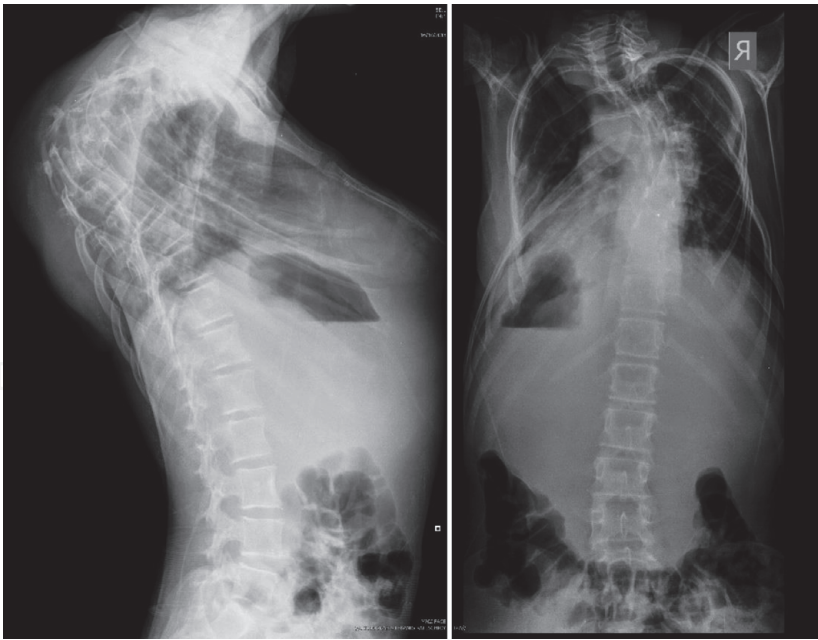
Spondylolisthesis is a rare complication in NF, caused from erosion or elongation of the pedicles or the pars intraarticularis, from neurofibroma. It requires stabilization in case of progression (**Figure 4**).



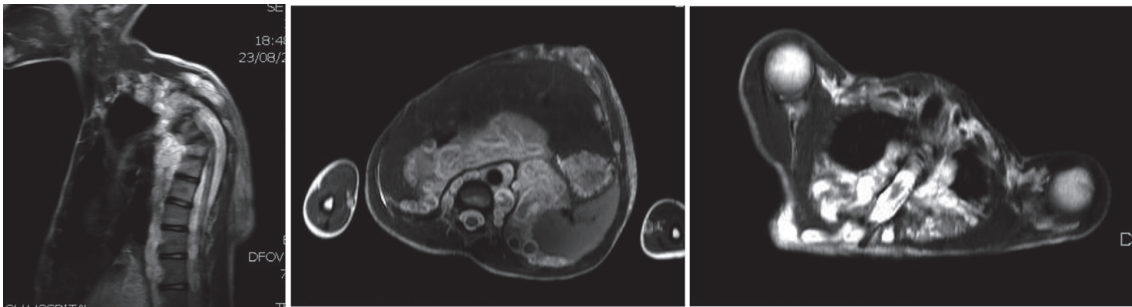
**Figure 4.**  
*Spondylolisthesis in a 12 yrs. old boy with NF-1, without signs of nerve compression. Observation and x-ray evaluation is recommended every year.*

## 2.4 Case presentation

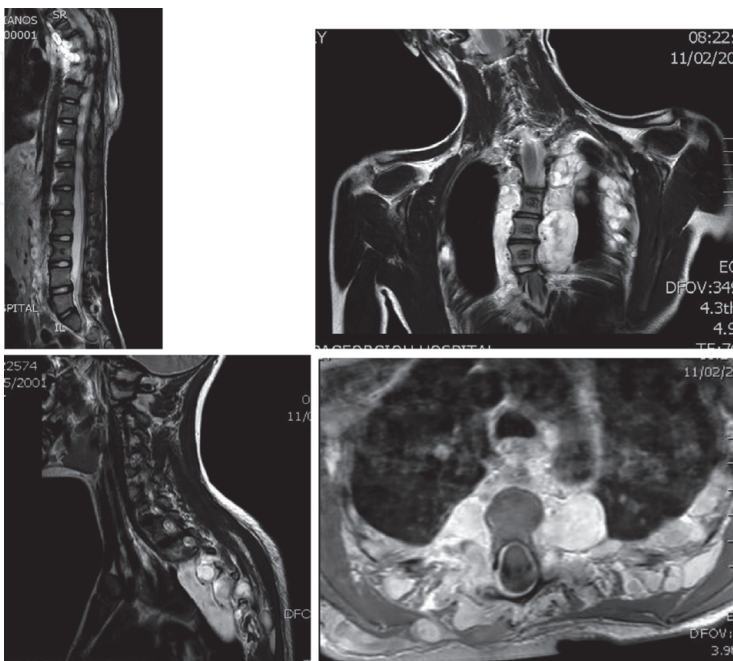
We present an adolescent boy with intrathoracic and intrabdominal neurofibromas that presented with dystrophic scoliosis and kyphosis and signs of paraplegia (**Figures 5–11**).



**Figure 5.**  
*Initial x-ray evaluation of upper thoracic dystrophic kyphoscoliosis, lateral and posterior view.*



**Figure 6.**  
*MRI abdomen and front spinal with the presence of intrathoracic paraspinal neurofibromas.*



**Figure 7.**  
*MRI of the upper thoracic spine with neurofibromas.*

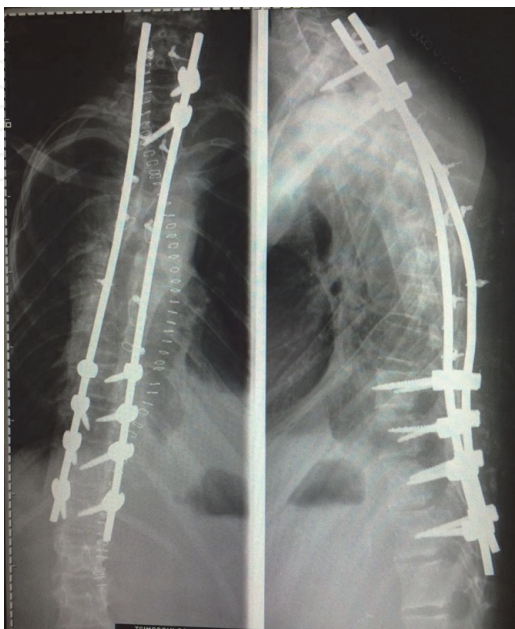




**Figure 8.**  
*Frontal and lateral view of the child with enlarged neurofibroma.*

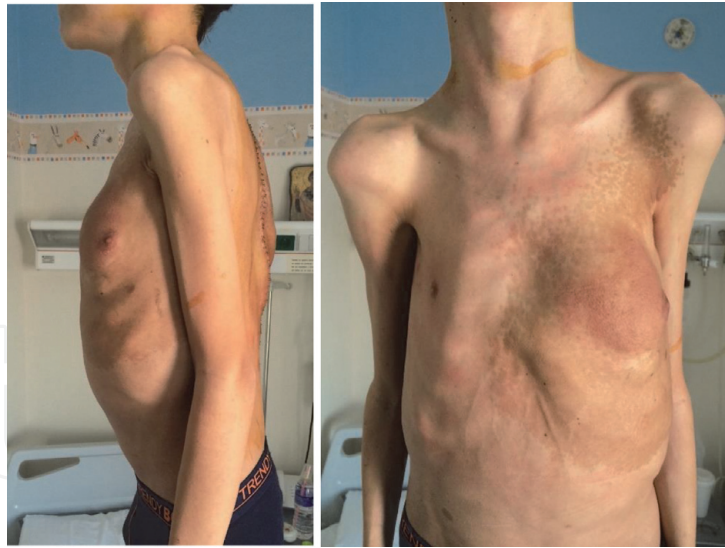


**Figure 9.**  
*CT evaluation with subluxation of the affected vertebrae.*



**Figure 10.**  
*After a period of 2 weeks in halo traction, surgical treatment with rods and pedicular screws. Surgeon: Vasileios Lykomitros. Spine surgeon, PhD, General Clinic Thessaloniki, Greece.*





**Figure 11.**  
*Impressive correction of the shape of the body.*

### 3. Osseous involvement of long bones

#### 3.1 Tibia

Lesions of long bones in NF patients are rare but create severe problems in their management.

The tibia is the most commonly affected long bone in NF. The incidence of congenital pseudoarthrosis of the tibia (CPT) is 1:250.000. It is most commonly associated with NF, but may appear with fibrous dysplasia or osteofibrous dysplasia.

The lesion appears with a characteristic anterior and lateral bowing, usually along with the presence of the skin manifestations of the disease. The lesion appears early in life; however, it must be distinguished from the congenital posteromedial bowing of the tibia. This has a benign course, with gradual improvement of the deformity, without underlying disease, usually leaving only problems of leg length discrepancy. Children affected with CPT have rarely simultaneously problems with scoliosis.

It may be associated with enlargement of the limb or with the presence of neurofibromas in the bone or in the surrounding tissue.

The posterior muscles of the calf are relaxed because of the anterior tibial bowing and the ankle is in a dorsiflexion position. The fibula is migrating proximally, leading in a valgus ankle position. Gradually leg length discrepancy (LLD) appears, due both to the tibial bowing and to altered function of the proximal and distal tibial growth plate. Hip alteration may appear in the form of coxa valga. This compensates the altered alignment of the tibia and there is overgrowth of the femur.

Radiological assessment of the tibia is the essential examination for evaluation of the severity of tibial involvement. The tibia is anteriorly and laterally bowed, most commonly in the distal third. The cortices present sclerosis with the presence of medullary canal, that may end without developing a fracture or pseudoarthrosis.

When cystic lesions appear with stenosis or loss of the medullary canal, the tibia becomes dysplastic and will present fracture or elements of pseudoarthrosis [19–26].

It is important to protect the limb since the anterior bowing is usually increasing gradually, leading either in fracture or forming the tibial pseudoarthrosis. Recently, guided growth of the bowed tibia with 8 plates, has been proposed, not only to prevent deformity and fracture but even to improve the axis of the tibia and fibula [27].

The fibula follows the bowing of the tibia but with smaller bowing, with increased thickness of the cortices as it carries more stress during walking. The ankle joint may be distorted, but the growth plate is not usually involved since the lesion is located in the diaphyseal lesion. There is alteration in the position of the growth plate that may be found in a recurvatum place. The distal part of the fibula is gradually migrating proximally [28].

Four radiologic types for CTP have been described from Crawford.

Type 1, non-dysplastic, with dense medullary canal. They have the best prognosis, that may end without a fracture.

Type 2 with an increased medullary canal and tubulation defect.

Type 3 with a cystic lesion. These patients require a surgical intervention before developing a fracture.

Type 4 Patients with fracture of the cyst that have developed pseudoarthrosis.

Recently Paley classified CTP in 4 types, each one with subtypes, taking in consideration both tibia and fibula [25].

Type 1 with anterolateral bowing of both tibia and fibula, without fracture.

Type 2 Fracture of the fibula, without tibial fracture, considering the possible fibular migration.

Type 3 Fracture of the tibia, without fibular fracture.

Type 4 Fracture both of tibia and fibula, considering 4 subtypes according to fibular migration and tibial bone defect.

The presence of neurofibromas in the area of the pseudoarthrosis are confirmed with MRI investigation, affecting the endosteal or periosteal area of the lesion.

The mechanism of development of pseudoarthrosis remains unknown. It may be the result of mechanical stress in the anterior bowing of the tibia, that has sclerotic cortices with eliminated canal. In the presence of neurofibroma in the endosteal, as it grows gradually, the cortices become thin and lead to fracture or pseudoarthrosis.

The periosteum is thickened, constricting the tibia and fibula, leading to atrophy. Resection of the thickened periosteum is part of treatment protocols used. Periosteal grafting has been used as a treatment option.

The osteoclastic activity of the periosteum is increased. The resorption of the bone graft is related to this increased osteoclastic activity. We have seen the delayed ossification of the proximal tibia, in the process of bone transport with the Ilizarov device. This can be explained by the generalised periosteum defect, for adequate bone formation [29, 30].

Treatment with bisphosphonates is now used to improve the bone formation in the pseudoarthrosis treatment. Stem cells harvested from the hamartoma tissue of CPT patients, have less osteogenic potential. We are using fibrin clot tissue, derived from the blood of the patients, to improve the union in surgically treated patients [31–33].

Neurofibromatosis lesions in the surrounding tissue interfere also with normal cortical development. In the histological specimen of the pseudoarthrosis tissue, removed at surgery, usually fibroblasts are found [19, 21].

Management of tibial pseudoarthrosis remains one of the most challenging issues, despite several methods that have been proposed. The strategy must target for the correction of the deformity, union of the pseudoarthrosis, restoration of the leg length discrepancy. Corrective osteotomies stabilized with plates and screws or

intramedullary devices present with a higher percentage of failure. The presence of neurofibromas in the surrounding tissues increase the surgical difficulties.

Microsurgery using the fibula for union of the pseudoarthrosis has been increasingly used in the last 20 years, improving the results of union [34, 35].

Early intervention, to prevent the development of pseudoarthrosis has better results. Prophylactic bypass grafting with an allograft fibula, has been reported in 10 patients, with no cases of tibial pseudoarthrosis [36].

Bone transport technique, after excision of the pathological bone specimen, can be used to restore the axis and the length discrepancy of the pseudoarthrotic tibia. The Ilizarov method has increased the rate of successful management of the tibial pseudoarthrosis [37, 38].

The use of Ilizarov external fixation can be combined with intramedullary rods to improve stability, with simultaneous iliac crest graft. Healing over the compression, with correction of LLD has been reported. The use of proliferative factors from stem cells are collaborative to achieve union [39–43].

Management of TCP with intramedullary rods alone in a cohort of 34 patients that reached skeletal maturity, was found to be functional in 82% of cases. Permanent IM rodding of the affected tibia is important factor for long term results [44].

Neurofibromas that are occasionally extending in large areas of the tibia, is difficult to be surgically removed. In these cases, amputation of the limb and use of prosthesis can be proposed [45, 46].

The cross-union concept initially presented from Choi et al. 2011 [47]. In children that both tibia and fibula were fractured, they converged the fibula ends to the tibia ends, creating a 4-in-1 bone osteosynthesis. They used cortical graft from the contralateral tibia and cortico-cancellous bone from the inner ilium table to form a layer posterior to the two bones, to complete the cross union of the tibia to the fibula. Paley [25] recently presented a 100% present union with this technique, combining the cross- union surgical technique with pharmacological agents. His protocol consists of presurgical administration of zoledronic acid. In surgery, removal of the hamartoma and the interosseous membrane. Rodding of both bones, the tibia with telescoping rods and fibula with wire. Application of a 3-layer graft from periosteum, cancellous bone and BMP2. Further stability provided initially with an Ilizarov device but recently with a locking plate.

### 3.1.1 Case presentation

We have treated a 15 years old adolescent with radical excision of the affected tibial pseudoarthrosis, with simultaneous bone transport from the proximal tibial.

We used the Ilizarov device to stabilize the bones and complete the bone transport. The affected tibia was extremely sclerotic. We removed 5 cm of bone, but the remaining ends were also sclerotic. There was an unexpected delay in the ossification process of the proximal part of the tibia, that was normal. After completing the docking of the transported bone, we removed the Ilizarov apparatus, performed an open procedure in the docking site, used bone graft and we performed plating of the docking site. We achieved the restoration of the LLD. Gradually the tibia appeared with bowing and x-ray examination revealed failure of union with loss of plating fixation. We repeated the osteosynthesis process with adequate reaming of the medullary canal, both proximal and distal, in very osteosclerotic cortices and inserted in the canal, the longitudinal half of the fibula. We augmented the graft that consisted from cortical and spongiosa from the ipsilateral fibula, with fibrin clot. We completed the operation with a revision of the plating. Fortunately, 2 years after the last operation the boy has signs of tibial union (**Figures 12–23**).



**Figure 12.**  
*Initial presentation antero medial bowing and sclerosis of the tibia in the x-ray.*

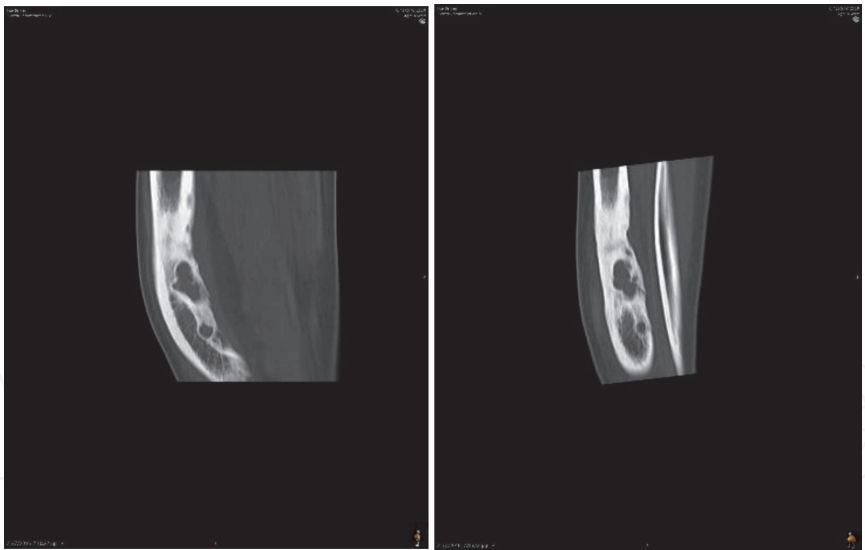


**Figure 13.**  
*The back of his mother, that has no osseous involvement.*

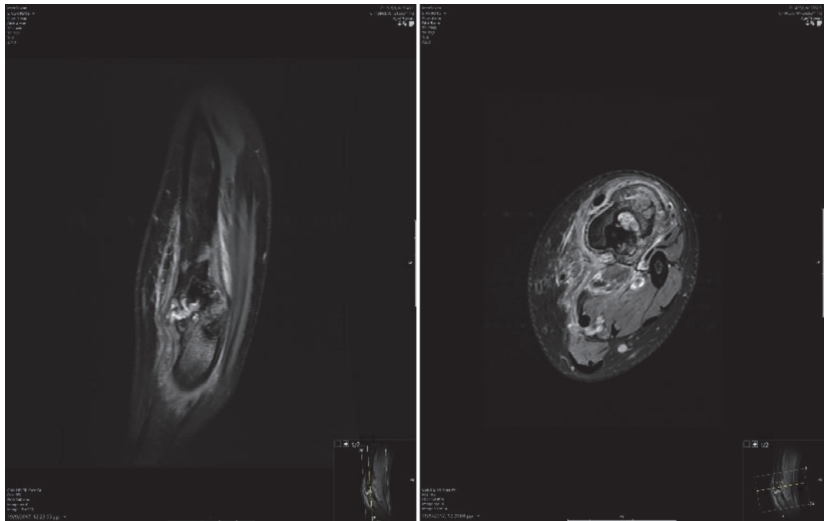


**Figure 14.**  
*X-ray of the tibia, with progression of the deformity and increased thickness of the fibula, with ankle malalignment.*

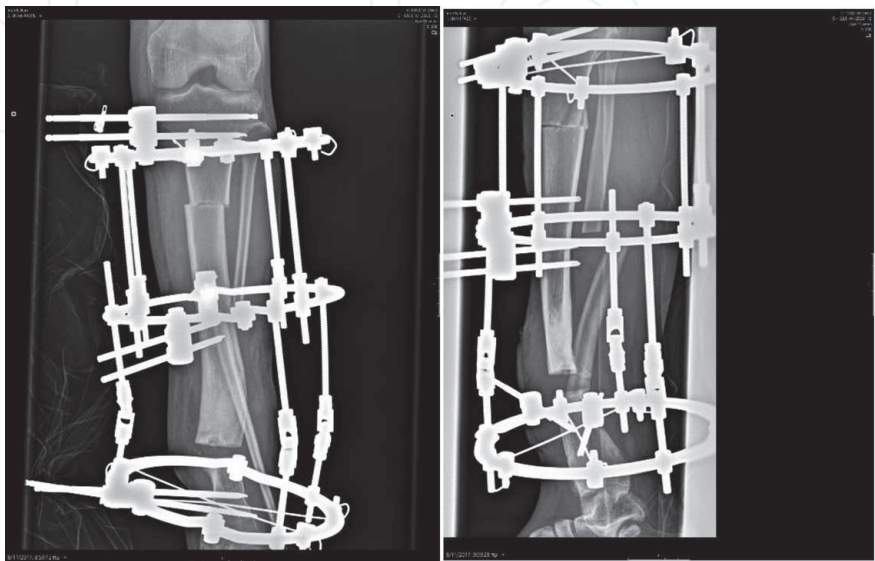




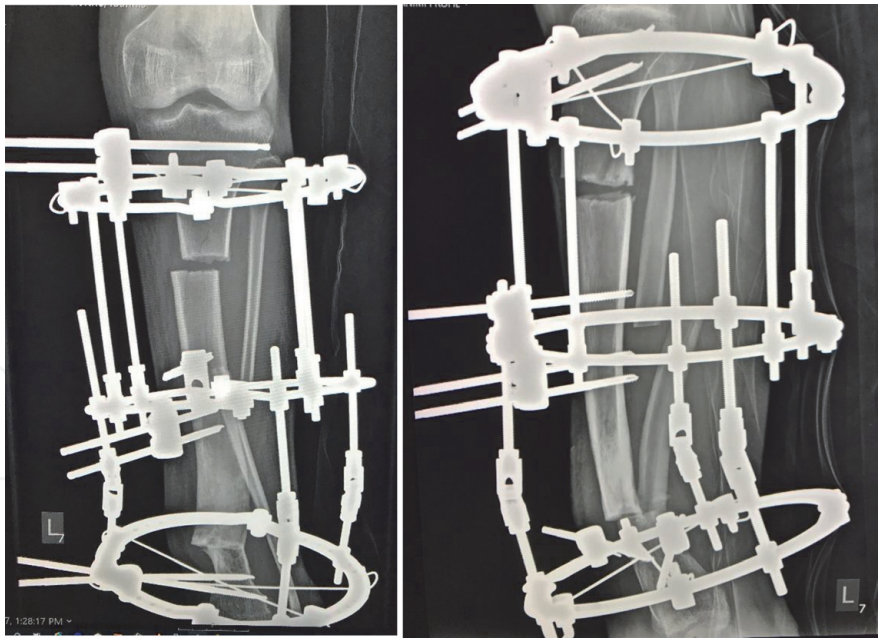
**Figure 15.**  
*CT scan of the cystic lesion of the tibia.*



**Figure 16.**  
*MRI of the pseudoarthrosis, with the presence of a diffuse neurofibroma in the area of the pseudoarthrosis.*



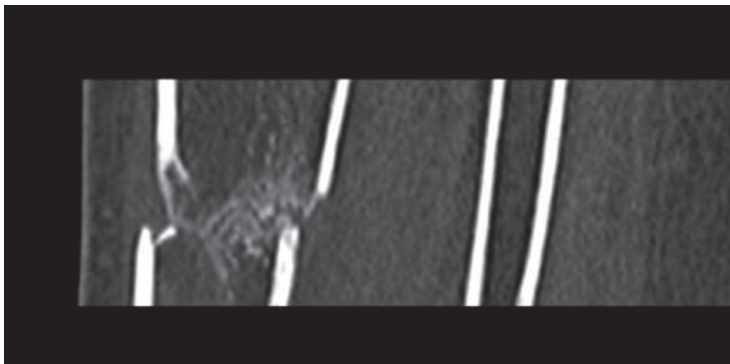
**Figure 17.**  
*Excision of the lesion, application of Ilizarov device with proximal osteotomy for bone transport. Simultaneous osteotomy of the fibula.*



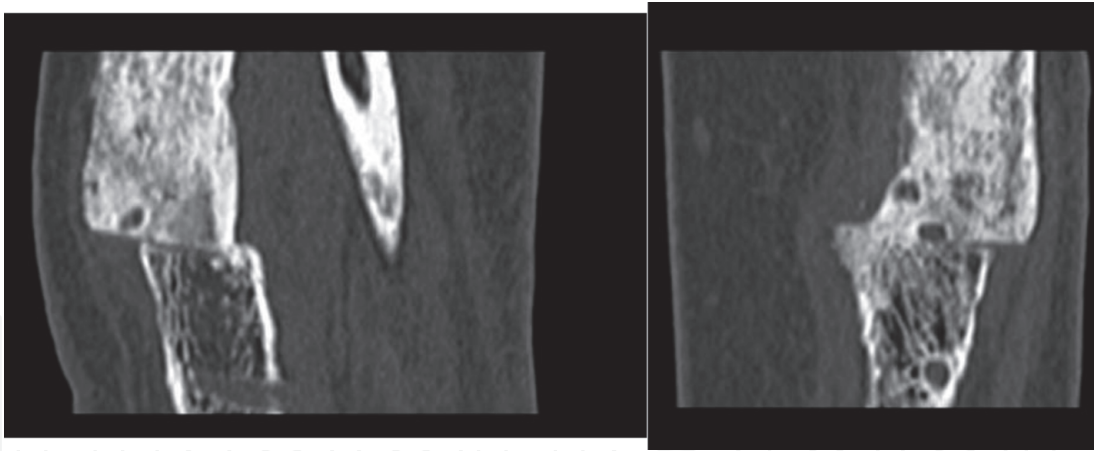
**Figure 18.**  
*At the end of the bone transport, at the docking day. Note the delayed ossification of the proximal osteotomy.*



**Figure 19.**  
*CT scan at the removal of the Ilizarov device.*



**Figure 20.**  
*CT scan with DELAYED OSSIFICATION at proximal osteotomy.*



**Figure 21.**  
*CT at the docking cite.*



**Figure 22.**  
*Mechanical failure of the plate, due to non-union of the tibial pseudoarthrosis.*



**Figure 23.**  
*Revision of the pseudoarthrosis, using the fibula as a strut in the tibial diaphysis, augmenting the procedure with bone graft and use of fibrin clot.*

### 3.2 Fibula

Congenital pseudoarthrosis of the fibula (CPF), isolated, is extremely rare, with few cases reported in the literature. It is highly associated with NF. Patients early in life present a valgus deformity of the ankle joint and an anterior bowing of the leg. There are other manifestations of the NF, usually the skin findings and the positive family history of NF. It may initially present with ankle varus deformity but as the fibula becomes pseudoarthrotic, it does not provide stability to the ankle. The fibula with the anterior bowing, migrates proximally and the talus is shifted in valgus position.

Dooley and Menelaus have classified CPF in four types.

Type 1: fibular bowing without fibular pseudoarthrosis, Type 2 fibular pseudoarthrosis without ankle deformity, type 3 with ankle deformity, and type 4 fibular pseudoarthrosis with late development of pseudoarthrosis of the tibia. It is important that the tibia appears with sclerosis of the medullary canal and the last type is a case of tibial pseudoarthrosis with fibular involvement, type 3 or 4 of the Crawford classification.

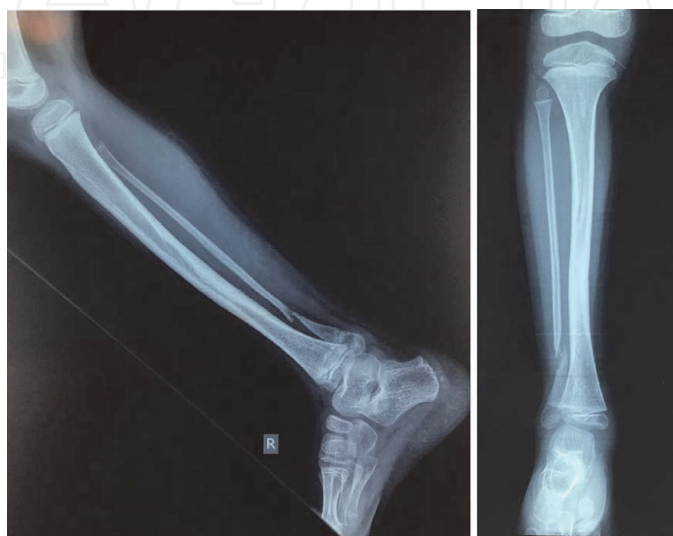
In the radiological examination, the fibula presents with pencilling of the pseudoarthrosis ends, with possible cystic formation, with anterior bowing. Distortion of the ankle joint, with valgus deviation result [48–51].

Ankle braces may be initially used to protect the ankle, but surgical management should be attempted early to protect the ankle joint. Initially Langeskiöld proposed the distal tibio-fibular fusion using bone graft. Treatment of the CPF aims to treat both the fibular pseudoarthrosis and the distortion of the ankle joint. Corrective osteotomies of the tibia and distal tibio fibular fusion, similar to Langeskiöld procedure are used. The Ilizarov apparatus is also used for treatment of the valgus deformity with distraction of the fibula and treatment of the pseudoarthrosis [52–55].

The use of periosteal flap from the diaphysis of the fibula and coverage of the pseudoarthrotic area is reported to have satisfactory results. The authors have treated 6 patients, after resection of the affected bone, retaining the proximal pedicle of the periosteum and suturing it as a tube in the defect [56].

#### 3.2.1 Fibula case presentation

We have treated our patient with thorough cleaning of the pseudoarthrosis site of the fibula. The histological specimen of the removed tissue revealed fibrotic



**Figure 24.**

*Initial x-ray presentation of fibula pseudoarthrosis. The talus has a valgus position, with proximal migration of the distal end of the fibula.*

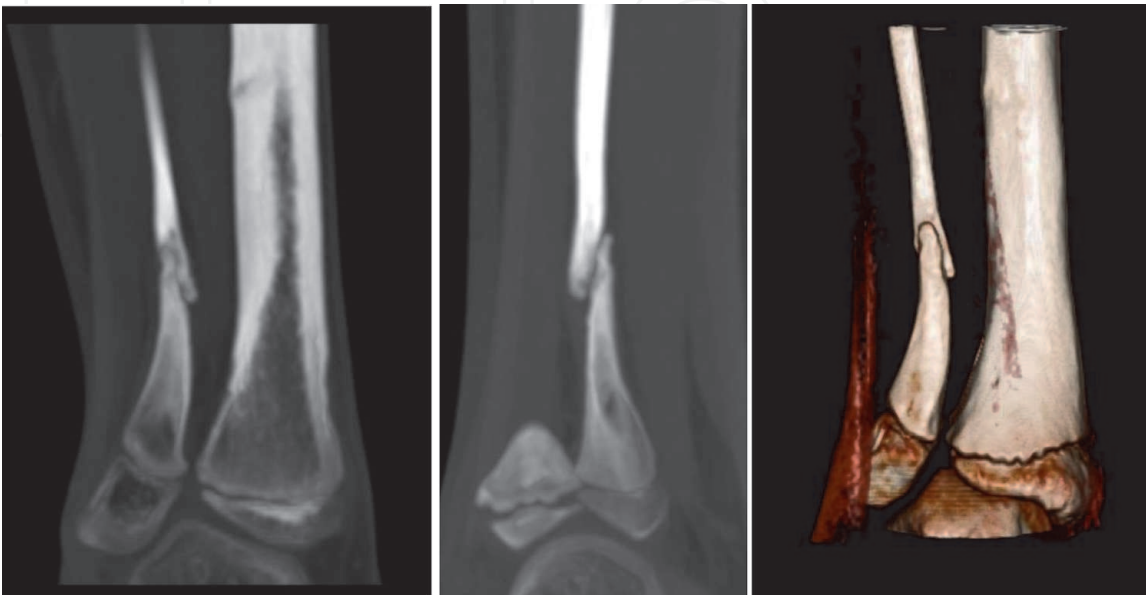




**Figure 25.**  
*Progression of the deformity, 2 years later.*



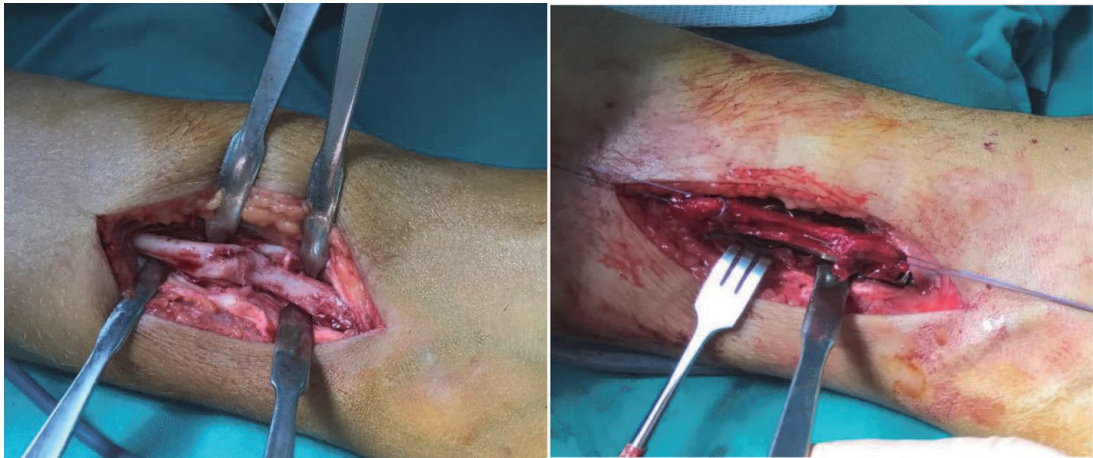
**Figure 26.**  
*Further increased valgus position of the ankle and more proximal migration of the distal part of the fibula, as the child grows.*



**Figure 27.**  
*CT scan of fibular pseudoarthrosis.*



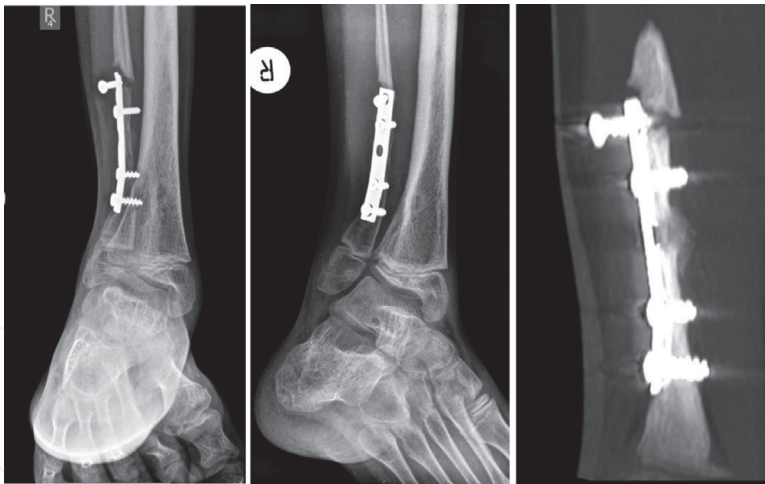
**Figure 28.**  
*Clinical picture of the valgus ankle right leg and the photo of the back of the patient with multiple café au lait spots.*



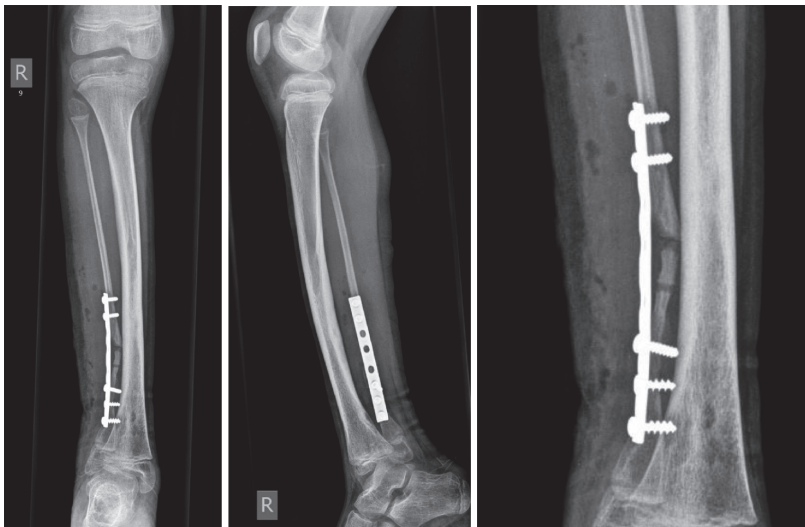
**Figure 29.**  
*Intraoperative picture of pseudoarthrosis of fibula and application of fibrin clot.*



**Figure 30.**  
*X-ray 3 months after surgery.*



**Figure 31.**  
*X-ray and CT scan, after failure of proximal screw and proximal fibular fracture. Pseudoarthrosis of the fibula is united.*



**Figure 32.**  
*X-ray, after revision of proximal fracture, with a longer plate. Pseudoarthrosis of the fibula is united.*

tissue. We stabilized the fibula using a semi tubular plate and adequate amount of bone graft with fibrin clot in order to enhance union. The fibula was extremely thin and despite that we use the fibular plate, the plate seems to be a little larger. In the latest examination, the patient is asymptomatic but despite that there was union of the pseudoarthrosis, the plate had failure in the proximal part and the fibula fractured in the area of the drill hole. We revised the plate, with a longer plate. We plan to improve the valgus position of the ankle, with 8 plate, as soon as the union will be complete (**Figures 24–32**).

### 3.3 Radius and ulna

Pseudoarthrosis of radius and ulna is an extremely rare condition, highly associated with NF. Patients present with a deformity of the arm, that is recognised early in life. It may present after an injury and failure to achieve union. The deformity is gradually increasing, despite that the function of the hand is not severely affected, children and parents are seeking for support. Radiological examination reveals the



presence of medullary sclerosis, cystic formation, obvious pseudoarthrosis, even type of agenesis of part of the affected bone.

Treatment of this condition is also very challenging. It requires the union of the pseudoarthrosis, restoration of the length of the affected bone and normal function of the wrist and elbow joint. As the child is growing, deformity is increasing, leading to distal radioulnar instability or to lesion of the radio capitellar joint. In ulnar pseudoarthrosis, with normal growth of the radius, the radial head will dislocate. All cases of untreated ulnar pseudoarthrosis ended with radial head dislocation.

Various surgical techniques have been used. Casting for treatment of pseudoarthrosis is not justified. Open reduction and stabilization with plate and screws with grafting has been reported to have a 23% success rate. The vascularized fibular graft with osteosynthesis has been referred to have the highest union rate [57–59]. The use of external fixator and Ilizarov technique is also reported. Either with excision of the affected part of the pseudoarthrotic bone, and bone transport, either with initial restoration of the length of the forearm and followed with vascularised fibular graft [60]. One bone arm treatment, with cross union of radius and ulna, has been proposed for the severe gap in the affected bones, having a high union rate but reducing the forearm function [61]. This is a salvage procedure.

Excision of the radius pseudoarthrosis, use of iliac crest graft, with shortening osteotomy of the ulna and stabilization of both forearm bones with intramedullary K wire was recently reported with sound union [62]. A double barrel vascularized fibular graft supplemented with k wires and external fixation, was used with success in a distal radius pseudoarthrosis [63].

### *3.3.1 Case presentation*

We present our patient who had ulnar pseudoarthrosis with dislocation of the radial head. She had almost normal use of the arm and elbow. Her parents had decided to avoid early surgery. The radial head had been protruding from the elbow, requiring surgical excision. The child was lost from our department (**Figures 33–35**).

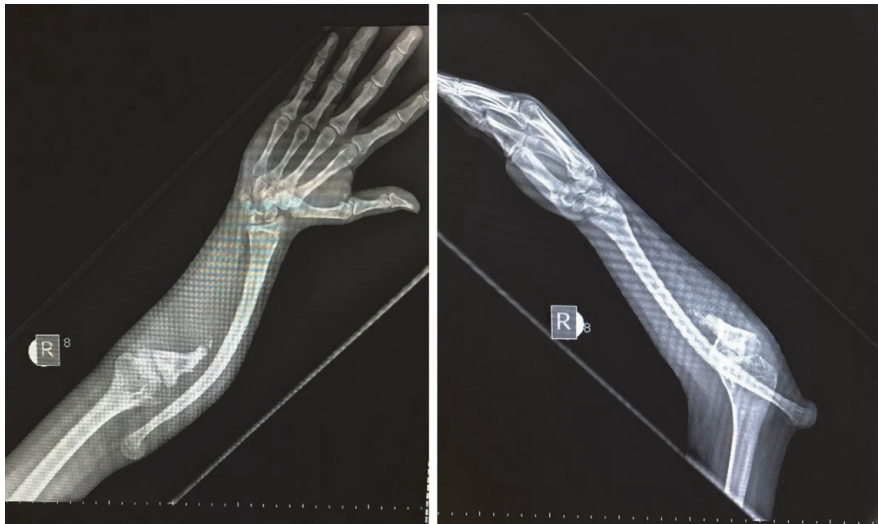


**Figure 33.**  
*Radiological presentation of ulnar pseudoarthrosis and progression to dislocation of the radial head.*





**Figure 34.**  
*The clinical picture, with the deviated short forearm, with radial head prominence, with almost normal function of the elbow.*



**Figure 35.**  
*Radiological picture of the dislocated radial head with the ulnar pseudoarthrosis.*

### 3.4 Overgrowth

The elephant man is the most known patient with neurofibromatosis. Overgrowth of the limb is usually associated with soft tissue enlargement, haemangiomas lesions or plexiform neuromas. These severe lesions are unilateral and associated with retroperitoneal fibromas that may require repeated surgery and possible may develop sarcomas. Surgical procedures of debulking have usually very limited successful results. The use of epiphysiodesis is an alternative to reduce the increasing leg length discrepancy. Crawford has described a patient that required hip disarticulation because of the tremendous overgrowth of the limb (Figure 36).



**Figure 36.**  
*Clinical picture of the overgrowth of the right leg, with café au lait spots in the leg and back of the child.*

#### 4. Conclusion

Children affected from neurofibromatosis must be regularly followed for osseous involvement. Examination of the spine, at least in a year basis, is important for the early diagnosis of spine deformities. Dystrophic curves are difficult to be managed conservatively. Improvement of radiological evaluation with MRI and CT and development of modern instrumentation permit us to manage effectively the progression of scoliotic curves.

Long bone involvement is one of the major clinical criteria for the diagnosis of NF-1. Treatment of congenital pseudoarthrosis remains one of the most complicated problems in children. Recent advances in biology and new implants have greatly improved our results.

#### Conflict of interest


The author declare no conflict of interest.

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